What is chronic myelomonocytic leukaemia?

Chronic myelomonocytic leukaemia (CMML) is a form of leukaemia characterised by high numbers of white blood cells called monocytes in the blood and bone marrow. It has been classified in the past as a form of myelodysplastic syndrome (MDS). The most recent classification schemes place CMML in a separate category.

About half of all patients have a form of CMML in which there is a high white cell count at diagnosis and the condition behaves most like a myeloproliferative neoplasm (MPN); the other half of patients have a normal or reduced white count at diagnosis and the disease behaves more like MDS. Experts do not agree on whether these are two different diseases or different stages of the same condition. Leukaemia & Lymphoma Research produce booklets on MPN and MDS.

CMML is mainly a disease of later life with the median age at onset being 70 years. A juvenile form of myelomonocytic leukaemia does occur (JMML), but this is not discussed in this factsheet. The incidence of CMML is about 1 to 2 cases per year per 100,000 population; men are more often affected than women.

What causes it?

The cause of CMML is unknown.

What are the signs & symptoms?

In both the MDS and the MPN types patients may complain of fatigue, weight loss, fever or night sweats. There is an increased risk of infections, and sometimes of bleeding due to low platelets. The spleen and/or liver may be enlarged in either type but this is more common in the MPN type (up to half of all patients).

How is it diagnosed?

The most important features in confirming the diagnosis are the blood count and bone marrow results. The disease is characterised by increased numbers of monocytes in the blood (>1x10^9/l) and in the marrow; fewer than 20% blasts (primitive leukaemic cells) in the marrow and an absence of the BCR/ABL genetic abnormality seen in CML. There are often chromosome abnormalities, and one rare abnormality may be associated with a response to a specific drug called Glivec.

How is it treated?

The MDS or non-proliferative type of CMML is treated in a similar fashion to MDS.

For most patients who are older, treatment is aimed at controlling the high numbers of
white blood cells. An oral chemotherapy drug called hydroxyurea should be the treatment of choice. Aggressive chemotherapy treatments (even including stem cell transplants), similar to those used for acute myeloid leukaemia (AML), may be recommended for younger patients but these are too toxic to be feasible for most CMML patients. A drug called Glivec, developed for treatment of chronic myeloid leukaemia (CML) may work for a very rare form of CMML in which patients have a specific chromosome abnormality (called t(5;12)). Patients with CMML may wish to discuss possible clinical trials with their specialist.

What is the prognosis?

Between 15 and 30% of cases of CMML will progress to acute myeloid leukaemia (AML). Leukaemia & Lymphoma Research produce a separate booklet on AML.

Median survival in CMML is around 20 months but ranges from about 10 months to over 5 years. There is no difference in survival between the MDS and MPN types of CMML. In a disease with such a variable prognosis it is very important for patients to discuss this with their specialist.

[Median Survival - a note
Median survival is often misunderstood by patients and family to mean the maximum expected lifespan. In fact, it is the time at which one would expect half of a group of patients diagnosed at the same time to still be alive - many of those still alive will live for many more years, decades even. It is also important to realize that not all patients who die after being diagnosed with CMML, die from CMML. Particularly in the case of elderly patients, many will die from other diseases. Finally, one should always remember that survival data is historical and may not reflect improvements based on newer drugs or treatments.]